

Necrotizing lymphadenitis: If not tuberculosis then what?

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ABSTRACT

We present a rare case of Kikuchi disease in a young lady presenting with fever and cervical lymphadenopathy. Clinically, the disease mimics tuberculosis and lymphoma. Lymph node biopsy is diagnostic and treatment is symptomatic. Although tuberculosis is endemic, clinicians should be aware of uncommon conditions as early recognition of the disease will minimize unnecessary evaluation and treatment.

Keywords: Kikuchi disease, necrotizing lymphadenitis, tuberculosis

Introduction

Kikuchi disease is a necrotizing lymphadenitis, is prevalent in Asia, and is being increasingly recognized in other areas of the world. It usually occurs in young women and is self-limited.^[1] The involvement of cervical lymph nodes with or without systemic manifestations is the most common presentation of Kikuchi disease. The initial clinicopathological appearance mimics lymphoma. It resolves spontaneously, usually over a period of several weeks to 6 months. It usually runs a benign course and recurrence is rare.^[2,3]

Case Report

A 31-year-old female presented with a 10-day history of high-grade fever and loss of appetite and generalized weakness. On examination, the patient had bilateral tender cervical lymphadenopathy in Level II and III.

Blood test revealed raised erythrocyte sedimentation rate and normal white blood cell count. Ultrasound of the neck showed

enlarged lymph nodes in Level II and III, largest size measuring about 2 cm. Chest X-ray was normal. Autoimmune and viral markers were negative.

Fine-needle aspiration cytology from the cervical node was suggestive of necrotizing lymphadenitis. For definitive diagnosis, biopsy was done which showed necrosis and the presence of crescentic histiocytes and plasmacytoid monocytes, suggestive of Kikuchi disease.

The patient was treated supportively and with steroids. The patient symptomatically improved and is on follow-up.

Discussion

Kikuchi disease also known as histiocytic necrotizing lymphadenitis was originally reported in young Japanese females in 1972 by Kikuchi and Fujimoto *et al.*^[4] The classic finding of Kikuchi disease is cervical lymphadenopathy and fever.^[5] Very few cases have been reported in India, and the first case was reported in 1998 by Mathew *et al.*^[6]

Most patients are young females below 40 years. The onset of disease is with fever and lymphadenopathy which is mostly cervical, in a previously healthy adult.

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The onset of Kikuchi disease could be acute or subacute, evolving over 2–3 weeks. The main clinical feature is unilateral lymphadenopathy, with cervical involvement in 70%–98% of cases. The jugular lymph nodes and posterior cervical chain are most commonly involved. Enlarged lymph nodes may vary in size and are tender.^[7] The involvement of mediastinal, peritoneal, and retroperitoneal regions are uncommon. Extranodal involvement of Kikuchi disease is rare and has been documented in the skin, bone marrow, myocardium, and central nervous system.^[8]

Important differential diagnosis of Kikuchi disease is tuberculosis, lymphoma, and systemic lupus erythematosus (SLE).

The definitive diagnosis of Kikuchi disease is made through lymph node excision biopsy and histologic examination. Antinuclear antibody is positive in about 7% of patients.^[9]

The etiology of Kikuchi disease is unknown; a viral, genetic, and an autoimmune hypothesis has been proposed. The role of Epstein–Barr virus, cytomegalovirus, and human herpesvirus-6 in eliciting a hyperimmune reaction leads by cytotoxic lymphocytes T toward infected lymphocytes has been emphasized. Among them, Epstein–Barr virus has been studied most extensively in Kikuchi disease, but no causal relationship has been demonstrated. Other investigators emphasize the role of immunological mechanisms involved in the pathogenesis of Kikuchi disease, related with SLE.^[10]

There is no specific treatment of Kikuchi disease. It usually resolves within 1–4 months. Analgesics, antipyretics, and nonsteroidal anti-inflammatory drugs can be used. Corticosteroids can be helpful in generalized disease and in aggressive clinical course.^[11]

Conclusion

Although the incidence of Kikuchi disease is rare, it must be considered among the differential diagnosis in young female patients presenting with fever and cervical lymphadenopathy.

Awareness among physicians about the disease is of crucial importance in minimizing unnecessary evaluation, misdiagnosis, and inappropriate treatment as the disease mimics tuberculosis and lymphoma.

In India, even though tuberculosis is rampant, not all cases of necrotizing lymphadenitis are tuberculosis. Hence, cautious use of empirical antitubercular therapy is recommended.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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